

SYSTEMIC SCLEROSIS: DIFFUSE AND LIMITED



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INTRODUCTION

The term scleroderma literally means hard skin. But scleroderma (also known as systemic sclerosis, or SSc) is much more than a skin disorder.

While nearly all persons with systemic sclerosis have issues with their skin, particularly excessive scarring, swollen, tight, or hard skin on their fingers, they also have organ system involvement. Many have tight, swollen, or hard skin in other body areas as well, particularly the face and the arms. Additionally, several other types of organ system involvement are also characteristic of systemic sclerosis, primarily the vascular and the immune systems. For example, blood vessels are frequently affected (vasculopathy), leading to spasmodic color changes (red, white, or blue) brought on by cold exposure. This is commonly referred to as Raynaud phenomenon and it occurs in the fingers of about 90 percent of persons with systemic sclerosis. One of the most common manifestations of immune involvement is the presence of abnormal levels of autoimmune antibodies to the nucleus of one's own cells (anti-nuclear antibodies or ANA) that are seen in nearly everyone with systemic sclerosis.

These three clinical features—excessive fibrosis (scarring), vasculopathy, and autoimmunity—appear to underlie the processes that result in the different manifestations that characterize systemic sclerosis.

TYPES OF SYSTEMIC SCLEROSIS

For reasons that are not clear, the normal protein collagen is deposited in the skin of patients with systemic sclerosis in higher amounts than in the skin of people who are disease-free. This process makes the skin thick and tough. In some persons, the excess collagen deposits involve only the fingers and possibly the face or hands. In others, the deposits can be found in areas all over the body. Furthermore, systemic sclerosis patients are placed into two additional subgroups based on the extent of skin involvement. These subgroups are diffuse cutaneous scleroderma and limited cutaneous scleroderma.

Diffuse cutaneous scleroderma

This subgroup is characterized by thick or tight skin on the arms, above and below the elbows, and frequently on the legs, above and below the knees, with or without involvement of the face. The skin on the torso (chest



and abdomen) is frequently tight, thick, or hard. The thickening of skin often progresses rapidly and is very bothersome, leading to thickening of skin in many areas all over the body in a short period of time (weeks to months). It is not uncommon for the thickening process in the skin in diffuse cutaneous scleroderma to continue for one to three years before the thickening process slows down and levels off. After one to two years of stability in the thickness, the thickening usually begins to recede and the skin begins to thin or soften.

Limited cutaneous scleroderma

People with this form of the disease have thick, tight, or hard skin on areas below, but not above, the elbows and knees, with or without involvement of the face. Thickening of skin frequently develops gradually and is relatively unobtrusive. When measured repeatedly over time, the skin score in patients with limited cutaneous scleroderma is usually small and changes very little, even over many years.

What does this division into limited and diffuse cutaneous scleroderma tell us?

Both subgroups are part of a more general disorder called systemic sclerosis and, as such, both subgroups

share some features in common, including:

- Raynaud Phenomenon. This occurs in about 90 percent of patients with systemic sclerosis.
- Heartburn and other esophagus problems (particularly trouble swallowing foods).
- Skin sores are common, primarily on the fingers. Some patients develop sores on the skin of the wrists, elbows, or ankles.
- Abdominal grumblings that can include feeling “full” after eating only a small amount of food, “bloating” of the belly after eating, swelling of the abdomen, particularly after eating, constipation, or diarrhea.
- About 10–15 percent of patients may develop severe lung fibrosis, leading to shortness of breath.
- About 10–20 percent of patients may develop pulmonary hypertension, which is high blood pressure in the arteries that supply the lungs.

While the above-listed characteristics are those that each subgroup has in common, there are features that are seen more commonly in one of the two subgroups.

Features seen more commonly in diffuse cutaneous systemic sclerosis:

- Kidney failure in about 15–20 percent of patients, fortunately for which there are treatments that can preserve kidney function and prolong life for many, if treated early.
- Approximately 10 percent of patients will experience some level of heart involvement, including fluid around the heart, heart rhythm disturbances symptomatic enough to require treatment, and possibly heart failure.
- Musculoskeletal aches and pains, decreased motion of some joints (fingers, wrists, elbows, shoulders, and occasionally knees) and declines in hand function that can result in disability are fairly frequent.

TYPICAL ORGAN INVOLVEMENT

Skin

The skin is the largest organ in the body. Its disadvantage is that it is spread over the surface of

the body and not located in one area, like the heart or the kidneys where it can be easily examined and measured. Because it is spread all over, new methods for measuring the degree of thickness over the body have had to be developed. The most frequently used assessment is a technique called skin scoring. Scoring is done by a medical professional who feels and pinches the skin in 17 body areas and assigns a number to the thickness of skin in each of the areas on a 0–3 scale (zero being normal and 3 being very thickened). The scores in all 17 areas are added together to give the skin score (the range of scores is 0–51). Using this technique, skin scores of patients with limited and diffuse cutaneous scleroderma have been documented to change over time in fairly characteristic ways.

Lungs

The vast majority of persons with systemic sclerosis will develop some scarring in the lungs, usually seen on a high-resolution chest computed tomography, or CATH scan. The majority of these persons will never develop serious lung involvement. However, in about 40 percent of patients, there will be some measurable decline in lung function. The vital capacity (a measure of how much air your lungs can move in and out in one deep breath) may get smaller because scars in the lung tissue make the lung stiffer. The good news is there have been encouraging treatment studies recently that suggest some immunosuppressive treatments (i.e. cyclophosphamide) may slow damage to the lungs. More research is ongoing that is looking into this and other treatment strategies to slow lung damage.

Kidneys

Kidney failure is a severe complication that occurs mainly in persons with diffuse cutaneous scleroderma who have had systemic sclerosis for less than five years. Problems begin with a decline in blood flow to the kidneys for reasons that doctors do not understand. This then triggers the release of hormones, which, unfortunately, results in elevations in blood pressure and further reductions in blood flow through the kidneys. This is commonly called

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- Helping patients and their families cope with scleroderma through mutual support groups, physician referrals and the National Patient Education Conference.
- Promoting public education of the disease through publications, seminars, patient education events and awareness activities.

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- Information and educational offerings from your local chapter.
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“renal crisis” and results in decreased kidney function, which doctors usually assess by measuring blood levels of creatinine, a normal breakdown product of human muscle.

Doctors have not yet developed ways to predict who will experience this complication. However, since doctors know that most patients who develop kidney failure also develop new onset high blood pressure at the same time, frequent blood pressure monitoring is prescribed. Patients are usually asked to obtain a home blood pressure monitoring device and start taking their blood pressures three times a week in an attempt to pick up the new onset of “renal crisis” as quickly as possible. If blood pressures go higher than 140/90 on two occasions 12 hours apart, patients are typically prescribed an angiotensin-converting enzyme, or “ACE” inhibitor. ACE-inhibitors are a class of medication that is especially tailored for use in this situation.

Unfortunately, even with these recommendations, some patients will go onto dialysis. Fortunately, more than half of the persons who start on dialysis will be able to get off dialysis usually within 18 months of onset of kidney failure as long as they continue taking ACE-inhibitors. If they cannot tolerate ACE-inhibitor treatment, they can and should be switched to an angiotensin-receptor blocker (like Cozaar or Diovan), even as they continue their dialysis.

Heart

When the kidneys fail, the heart frequently fails—at least temporarily. When the kidneys come under control, the heart usually returns to normal. Unfortunately a small percentage of patients will have scars in the heart muscle that will be the cause of abnormal heart function. Doctors have not yet come up with good treatments for this kind of heart problem.

Gastrointestinal

About 80–90 percent of persons with systemic sclerosis can be shown to have a “lazy” muscle in their esophagus. This can lead to heartburn and the sensation that food “sticks” in the chest part way

down. Unfortunately, the muscle in the stomach and in the rest of the intestine can also get lazy. This can lead to a multitude of symptoms including filling up too fast after eating small amounts of food, a sensation of “bloating” or swelling in the belly after eating, chronic diarrhea or chronic constipation. Unfortunately many of these symptoms are not specific to any particular area of the gut and it may take a doctor and patient considerable time to try to sort out which area of the gut is causing the problem and which area needs treatment.

Musculoskeletal Pain

The majority of persons with systemic sclerosis also experience musculoskeletal pain of some sort, particularly in the first few years. Arthritis in the joints, inflammation in the tendons (tendonitis) and bursa (bursitis), diffuse myalgias (muscle pains), or fibromyalgia (aches and pains all over the body, accompanied by soft tissue areas which are tender to mild palpation), are common. Early on, patients with diffuse cutaneous scleroderma are more likely to have musculoskeletal pain that often begins to ease when the skin starts to soften.

PREDICTING THE FUTURE

Doctors can learn much about the future of systemic sclerosis patients during the first years of the disease. The majority of negative health outcomes that can happen in systemic sclerosis will have happened in the first five years or they are not likely to occur at all. So, if significant heart, lung, or kidney problems have not occurred within that time, it is unlikely they will begin anew after five years. In general, by the fifth year, the pace of disease activity will slow down, the skin will begin to soften, and the fatigue, musculoskeletal aching, and joint motion will improve.

Please note that this brochure is provided for educational purposes only. It is not intended to substitute for informed medical advice.

The Scleroderma Foundation wishes to thank Philip Clements, M.D. for his input on this brochure.

OUR THREE-FOLD MISSION IS SUPPORT, EDUCATION AND RESEARCH

Support: To help patients and their families cope with scleroderma through mutual support programs, peer counseling, physician referrals, and educational information.

Education: To promote public awareness and education through patient and health professional seminars, literature, and publicity campaigns.

Research: To stimulate and support research to improve treatment and ultimately find the cause of and cure for scleroderma and related diseases.



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